

IN THE CLAIMS:

1-20. (Cancelled).

21. (Currently Amended). A method of slowing mucopolysaccharidosis disease progression in a patient in need thereof consisting of comprising administering an inhibitor of glucosylceramide synthase in an efficacious amount to slow mucopolysaccharidosis disease progression in a patient, wherein the inhibitor of glucosylceramide synthase is N-butyldeoxynojirimycin.

22. (Currently Amended). The method according to claim 21 wherein the mucopolysaccharidosis disease is selected from the group consisting of MPS I (MPS IH, IS or IH/S), MPS II, MPS IIIA, IIIB, IIIC or IIID, MPS IVA or IVB, MPS VI and MPS VII.

23-27. (Canceled).

28. (Currently Amended). A method of reducing neuronal glycolipid storage in mucopolysaccharidosis disease in a patient in need thereof consisting of comprising administering an inhibitor of glucosylceramide synthase in an efficacious amount to reduce neuronal glycolipid storage in mucopolysaccharidosis disease in a patient, wherein the inhibitor of glucosylceramide synthase is N-butyldeoxynojirimycin.

29. (Currently Amended). The method according to claim 28 wherein the mucopolysaccharidosis disease is selected from the group consisting of MPS I (MPS IH, IS or IH/S), MPS II, MPS IIIA, IIIB, IIIC or IIID, MPS IVA or IVB, MPS VI and MPS VII.

30-35. (Canceled).

36. (Currently Amended). A method of reducing pathological features resulting from glycolipid accumulation in a patient with a mucopolysaccharidosis disease consisting of comprising administering an inhibitor of glucosylceramide synthase in an efficacious amount to reduce pathological features resulting from glycolipid accumulation in a patient with a mucopolysaccharidosis disease, wherein the inhibitor of glucosylceramide synthase is N-butyldeoxynojirimycin.

37. (Currently Amended). A method for improving survival of a patient with a mucopolysaccharidosis disease consisting of comprising administering an inhibitor of glucosylceramide synthase in an efficacious amount to improve survival of a patient with a mucopolysaccharidosis disease, wherein the inhibitor of glucosylceramide synthase is N-butyldeoxynojirimycin.

38. (Currently Amended). A method of slowing mucopolysaccharidosis disease progression in a patient in need thereof consisting of comprising administering an inhibitor of glucosylceramide synthase in an efficacious amount to slow mucopolysaccharidosis disease progression in a patient, wherein the inhibitor of glucosylceramide synthase is N-butyldeoxygalactonojirimycin.

39. (Currently Amended). The method according to claim 38, wherein the mucopolysaccharidosis disease is selected from the group consisting of MPS I (MPS IH, IS or IH/S), MPS II, MPS IIIA, IIIB, IIIC or IIID, MPS IVA or IVB, MPS VI and MPS VII.